452 Trials, tribulations and triumphs of a cystic fibrosis study – a behind the scenes look at the workings of an international multi-centre study

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Background: Randomized controlled trials (RCT) are an essential part of health care as we search for evidence-based outcomes to ensure best clinical practice. Research results are reported and evaluated; however, the process of research is seldom reviewed.
Objective: To describe some of the difficulties and achievements in conducting a multi-centre research study over an extended period of time.
Methods: The Australasian Cystic Fibrosis Bronchoalveolar Lavage (ACFBAL) Study ran over 10 yrs in 8 sites throughout Australia and New Zealand. Prospective data were collected for this RCT designed to examine BAL directed therapy in children diagnosed on newborn screening. 168 infants were randomized at <6 months. Routine clinical management as well as exacerbations and hospitalizations were recorded for each patient. A standardized protocol was used across all sites.
Results: Difficulties: Ethics approval across all sites took 4 yrs. The pharmaceutical company changed hands 3 times; there were 4 data managers and at least 2 co-ordinators at each of the 8 sites. Legal contracts replaced handshakes and quick emails.
Achievements: An amazing 93% of patients completed their 5 yrs on the study. Over 1000 admissions, 2000 exacerbations, 2800 reviews, 2700 routine checks, 500 BALs, 1800 cough specimens, and 400 audiology tests were recorded. More than 350 blood and 1000 BAL specimens were moved to sites in Australia and overseas. None went astray or deteriorated in transit.
Conclusions: It is possible to have long term, long distance relationships that have happy endings but all parties need to be committed to the relationship!
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453 Prevalence of CF-related chronic rhinosinusitis – results from a multicentre interdisciplinary study

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Rationale: The paranasal sinuses are regularly involved in CF but the prevalence of chronic and intermittent rhinosinusitic symptoms is discussed controversially. As an example, the reported frequency of smelling disorders in CF varies between 12 and 71%.
Methods: 187 CF-patients of all ages from 5 centres were assessed for sinonasal symptoms in the preceding year by questionnaire. EPOS-criteria (European Position Paper on Rhinosinusitis and Nasal Polyps 2007) were applied to quote the prevalence of chronic and intermittent rhinosinusitis.
Results: 31% of the CF-patients fulfilled the EPOS-criteria of chronic rhinosinusitis and 34% suffered from remittent ENT problems. We present frequency of the single symptoms. In 10 of 187 patients (5.3%) sinonasal symptoms were the cardinal symptom leading to diagnose of CF.
Discussion: Whereas about 100% of CF-patients have morpholgical changes in upper airway CT (Eggesbo 2002) and 93% revealed pathological changes in nasal cytology (Babinski 2008) each 1/3 of patients suffer form intermittent respective chronic rhinosinusitis. Remarkably, Glicklich (1995) et al. showed that the health impact of these symptoms could be greater than currently appreciated: patients with chronic sinusitis revealed a significantly higher burden of their symptoms in bodily pain and social functioning compared with chronic diseases like congestive heart failure and COPD.
Conclusions: Upper airway involvement in CF is underestimated and it requires further scientific and clinical attention.

454 Survey on therapy of CF-related chronic rhinosinusitis – results from a multicentre interdisciplinary study

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Rationale: As impairment of mucociliar clearance in CF also concerns the upper airways, almost all patients have pathological sinonasal changes. This restrains quality of life and it can accelerate progression in CF e.g. by first infection and persistence of *P. aeruginosa* in the sinuses. Previously, we showed in our interdisciplinary multicentre study including 187 CF-patients of all ages that CF-patients with *P. aeruginosa*-colonization of both airway segments regularly harboured genotypically identical strains in the upper and lower airways (95.8%).
Methods: Within the same study we assessed the surgical and conservative therapeutic measures for rhinosinusitis symptoms taken in the participating CF-centres.
Results: The fraction of CF-patients who underwent ENT-surgery differed substantially among the 3 major recruiting centres (30.4%, resp. 48.4% and 62.1%) and patients underwent up to 5 surgical interventions for high recurrence rates. Regarding conservative therapy 12/187 patients reported to abuse alfa-sympathomimetics. Nasal douches for removement of mucus and crusts only were utilized by 18.4% of patients and the applied volumes varied from 10–1000 ML a douche.
Discussion: The huge variance in surgical and conservative therapeutic measures demonstrates the lack of evaluated therapeutic concepts in the upper airways' involvement. Further research on its background, diagnostics and therapy are required with the aim to establish interdisciplinary concepts of care for this manifestation.

455 Paranasal sinus mucoceles in children with cystic fibrosis

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Introduction: Recurrent and chronic rhinosinusitis and nasal polyposis represent typical otolaryngological manifestations in patients with cystic fibrosis (CF). Recently, some CF-children with paranasal sinus mucocele have been described. The authors reported their experience about this pathological appearance focusing on diagnosis and treatment of such dangerous feature.
Material and Methods: A cohort of 15 cases of paranasal sinus mucocele in CF-children was managed. Ten patients were male and 5 females, with a mean age of 5 years (13 mo–18 ys). Preoperative work-up included nasal endoscopy and CT of sinus. All children underwent endoscopic nasal surgical treatment.
Results: CT showed a maxillary, ethmoid, and sphenoid mucocele in 12, 3, and 2 patients, respectively. Bilateral mucoceles were observed in 11 children and 3 patients presented mucoceles involved different paranasal sinuses. Surgeries were performed in presence of bone erosion, and/or esophtalm, and/or nocturnal desaturation.
No perioperative complications were evident. To date, no recurrences have been observed after a mean follow-up of 33 mo (3 mo–5 yr).
Conclusions: Cystic fibrosis can be considered a predisposing factor for the occurrence of paranasal sinus mucocele. To the best our knowledge, this is the largest series of CF-children with paranasal sinus mucocele. Preoperative CT is mandatory to assess this paranasal disorder. Endoscopic surgical treatment is safe, effective, and less invasive. A close collaboration with otolaryngologist is fundamental to obtain successful results.